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【 CASE REPORT 】

Clinical Utility of Diffusion-weighted Whole-body Magnetic Resonance Imaging with Background Body Signal Suppression for Assessing and Monitoring IgG4-related Disease

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Abstract:

A 77-year-old man with symptoms of chest pain was diagnosed with immunoglobulin G4 (IgG4)-related disease. Fluorodeoxyglucose positron emission tomography/computed tomography (FDG-PET/CT) revealed an intense uptake in the submandibular gland, lymph nodes and abdominal aortic wall. Diffusion-weighted imaging with background body signal suppression (DWIBS) revealed signal enhancements at the same location as those of the FDG-PET/CT findings. The DWIBS signal intensity decreased after steroid treatment, so we decreased the steroid dosage. Relapse did not occur. DWIBS makes it possible to adjust the medicine dosage while confirming the therapeutic effects and will likely be a useful method for monitoring IgG4-related disease.

Key words: IgG4-related disease, DWIBS, MRI

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Introduction

Immunoglobulin G4 (IgG4)-related disease, which can affect multiple organ systems, including the endocrine, urinary, and cardiovascular systems, is being increasingly recognized (1). This disease is diagnosed by the presence of a mass or swelling of organs, elevated serum IgG4 levels, and prominent IgG4-positive plasmacyte infiltration of involved organs (2).

Some reports have suggested that fluorodeoxyglucose positron emission tomography/computed tomography (FDG-PET/CT) may be useful for assessing the organ involvement and monitoring the therapeutic response in IgG4-related disease (3, 4). However, FDG-PET/CT has some issues, including elevated radiation exposure and a high cost. Diffusion-weighted imaging with background body signal suppression (DWIBS), introduced by Takahara et al. (5) in 2004, is a relatively new imaging modality. It is primarily used for cancer screening or staging, and a few reports have addressed the utility of DWIBS for patients with arteritis (6, 7).

We herein report our assessment of the organ involvement and monitoring of the therapeutic response in a case of IgG 4-related disease using DWIBS.

Case Report

A 77-year-old man underwent emergency invasive coronary angiography (CAG) for unstable angina. CAG showed severe multivessel disease. We suspected IgG4-related disease from intravascular ultrasound (IVUS) findings and performed further tests. He had a high serum IgG4 level and abundant IgG4-positve plasmacyte infiltration on lymph node biopsy specimens (8).

Fluorodeoxyglucose positron emission tomography/computed tomography (FDG-PET/CT) demonstrated an intense uptake in the submandibular gland, lymph node, and ab-

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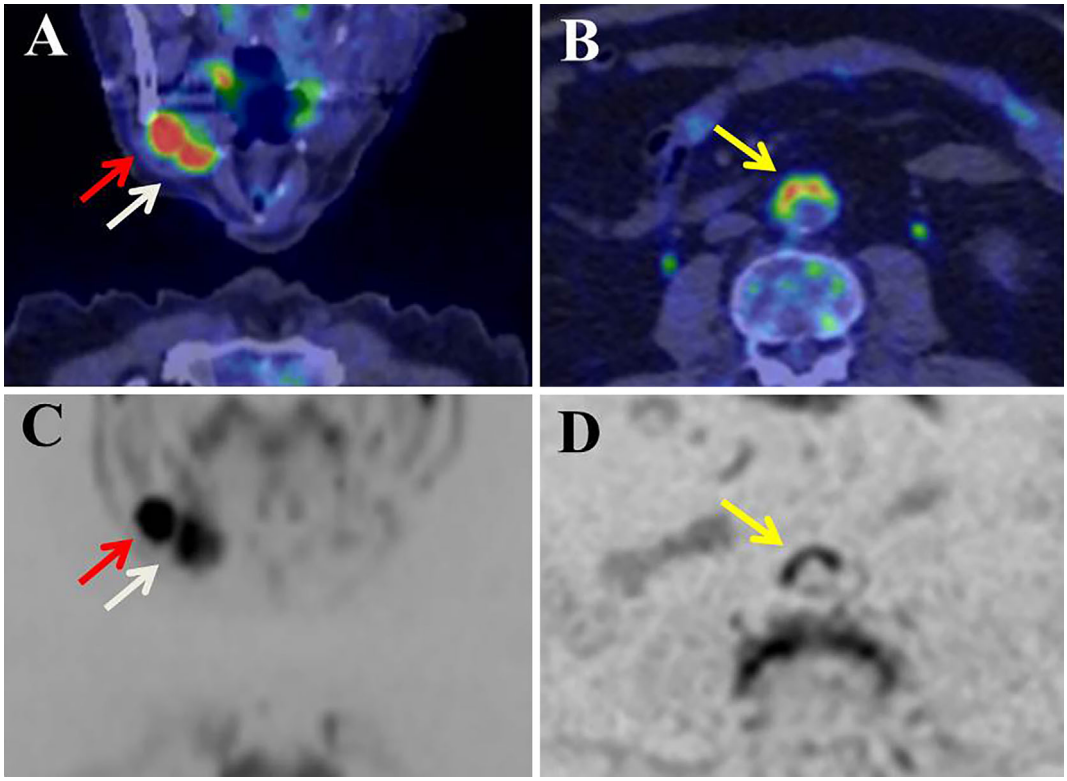

Figure 1. **(A) Coronal image of FDG-PET/CT before the treatment. The accumulation of FDG is observed in the submandibular gland (white arrow) and lymph nodes (red arrow). (B) Axial image of FDG-PET/CT before the treatment. The accumulation of FDG is observed in the abdominal aorta (yellow arrow). (C) Coronal image of DWIBS before the treatment. High signals are also observed in the submandibular gland (white arrow) and lymph nodes (red arrow). (D) Axial image of DWIBS before the treatment. High signals are observed in the abdominal aorta as well (yellow arrow).**

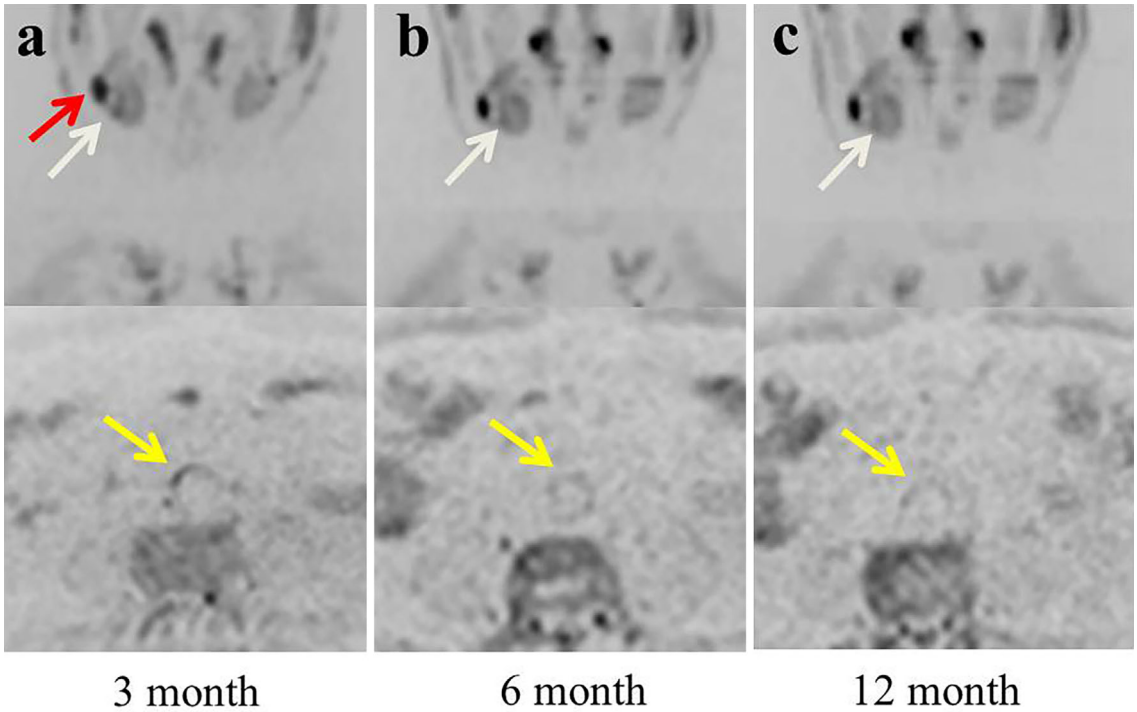

Figure 2. **(a) The high signals from the submandibular gland (white arrow), lymph nodes (red arrow), and abdominal aorta (yellow arrow) decreased after 3 months' treatment according to DWIBS. (b) The signal from the submandibular gland (white arrow) and abdominal aorta (yellow arrow) had further decreased after 6 months' treatment according to DWIBS. (c) The decline in the signals persisted after 12 months' treatment according to DWIBS.**

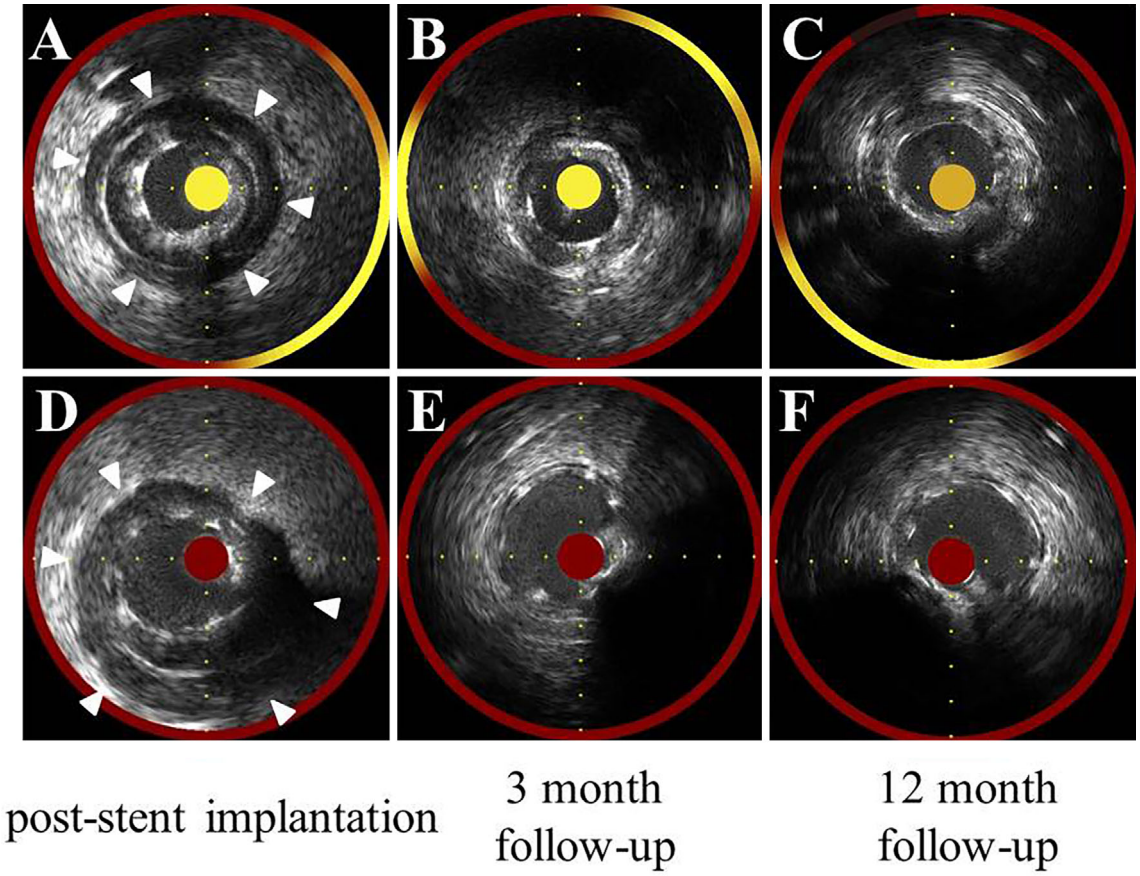

Figure 3. **(A) Near-infrared spectroscopy intravascular ultrasound (NIRS-IVUS) imaging immediately after stent implantation of the left circumflex artery (LCX) showed a low-echoic region on the outer side of the media (white arrowheads). (B) NIRS-IVUS imaging 3 months after treatment of the LCX showed a markedly reduced low-echoic region on the outer side of media. (C) The reduced low-echoic region persisted after 12 months' treatment according to NIRS-IVUS imaging. (D) NIRS-IVUS imaging immediately after stent implantation of the right coronary artery (RCA) showed a low-echoic region on the outer side of the media (white arrowheads). (E) NIRS-IVUS imaging 3 months after treatment of the RCA showed a markedly reduced low-echoic region on the outer side of media. (F) The reduced low-echoic region persisted after 12 months of treatment according to NIRS-IVUS imaging.**

dominal aortic wall (Fig. 1A, B). We performed DWIBS around the same time, which also revealed signal enhancement of the submandibular gland, lymph node, and abdominal aortic wall (Fig. 1C, D).

He complained of thirst due to reduced salivation. We therefore decided to treat him with an oral steroid. Oral steroid therapy was started with prednisolone at 40 mg per day before being tapered to 20 mg for 6 weeks. His complaints of thirst improved after prednisolone treatment. During 1-year follow-up with steroid therapy, the serum IgG4 levels decreased from 773 mg/dL to 51 mg/dL, and the soluble interleukin-2 receptor levels also decreased from 902 U/mL to 182 U/mL. By month 3, the high signals from the submandibular gland, lymph node, and abdominal aortic wall had decreased on DWIBS. These signals had decreased further by month 6 and 12 (Fig. 2). We also confirmed marked shrinking of the pseudotumors surrounding 2 coronary arteries on IVUS findings at months 3 and 12 (Fig. 3). We were able to reduce the prednisolone dose to 5 mg/day by 12 months based on DWIBS findings without any relapse.

Discussion

IgG4-related disease can affect multiple organ systems, including the endocrine, urinary, and cardiovascular systems. IgG4-related cardiovascular disease mainly involves the aorta, and IgG4-related coronary artery disease remains uncommon (9). Recently, several cases have been reported that involved IgG4-related coronary disease. Some patients had pseudotumors surrounding coronary arteries (10-14), and others had aneurysms of coronary arteries (15-20). The present patient had pseudotumors surrounding two coronary arteries with stenosis. In general, steroids are effective for treating coronary pseudotumors and lead to a marked reduction in the size of the mass lesions around the arteries. However, if patients have an aortic aneurysm, physicians

should be aware of the risk of bursting when administering large amounts of steroids. Indeed, IgG4-related aneurysm rupture has occasionally been reported (21, 22). High-dose corticosteroids might suppress inflammation and prevent further IgG4-related aneurysm development, but it may also increase the risk of rupture of developed aneurysms by making the arterial wall thinner. The present case did not have any aneurysm, and the mass lesion around coronary arteries was thus successfully reduced after steroid treatment.

FDG-PET/CT has been reported to be useful for the diagnosis and treatment of IgG4-related disease (23). FDG-PET/CT enables the acquisition of whole-body images and provides functional information on the disease activity. The disadvantages of FDG-PET/CT, however, are a relatively long preparation time for the examination and exposure of the patient and examiner to ionizing radiation. In addition, in order to obtain good-quality images, adjusting the concentration of circulating glucose is essential, which can significantly affect the FDG uptake.

Magnetic resonance imaging (MRI) does not have these disadvantages and is able to provide both anatomical and functional information within a single examination. Whole-body DWIBS, which was introduced by Takahara et al. (5) in 2004, has been used increasingly frequently, and many studies have confirmed that it is a feasible clinical technique for assessing the original and metastatic lesions of adult patients with high sensitivity and accuracy (24, 25). In many pathological conditions, water diffusivity is impeded due to increased neoplastic cellularity and swelling in inflammatory or infectious lesions. These conditions can be detected with DWIBS. We tried to perform DWIBS for the present patient with IgG4-related disease. This modality also revealed the signal enhancement of the tissues, similar to the FDG-PET/CT findings. DWIBS is an excellent tool for evaluating the disease activity in cases of IgG4-related disease; however, there is a limitation. Since DWIBS has difficulty depicting certain areas because of signal loss due to heart motion, it cannot be used to evaluate the heart. We must be aware of these limitations of DWIBS when using it to evaluate cases of IgG4-related disease.

Advantages of DWIBS include the avoidance of ionizing radiation and a low cost compared to FDG-PET/CT. These advantages allow DWIBS to be used for therapeutic monitoring. The effects of steroid treatment were evaluated by DWIBS at months 3, 6, and 12 in the present case. The signals of the submandibular gland appeared to decrease more rapidly than those of the abdominal aortic wall in our patient. There may be tissue-specific differences in the effects of steroid treatment; however, these points will need to be explored in further studies.

In summary, we herein report the first successful use of DWIBS for the evaluation and follow-up of IgG4-related disease. Since DWIBS is non-invasive, it can be performed repeatedly. Using this modality, it is possible to adjust the amount of medication administered while confirming the therapeutic effect, making it highly likely that DWIBS will be recognized as a useful test in the future.

The authors state that they have no Conflict of Interest (COI).

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